Acta Neurol Scand 1993: 87: 505–509
Printed in Belgium – all rights reserved

ACTA NEUROLOGICA SCANDINAVICA
ISSN 0001-6314

Post-traumatic segmental myoclonus associated with bilateral olivary hypertrophy

Birbamer G, Gerstenbrand F, Kofler M, Buchberger W, Felber S, Aichner F. Post-traumatic segmental myoclonus associated with bilateral olivary hypertrophy.
Acta Neurol Scand 1993: 87: 505–509. © Munksgaard 1993.

We present clinical and magnetic resonance (MR) findings in three patients with segmental myoclonia occurring 11–18 months after severe brainstem injury. Palatal myoclonus and vertical ocular myorhythmia were present in all three patients and synchronous involuntary movements of the upper extremities ("wing beating") in two patients. MR-imaging showed multiple post-traumatic lesions within the dentato-rubro-olivary pathway ("myoclonic triangle"), associated with bilateral enlargement and increased signal intensity of the inferior olives. The signal abnormality was more prominent on proton density weighted images than on T2-weighted images, suggesting underlying pathological changes different from typical gliosis.

G. Birbamer [1,2], F. Gerstenbrand [2], M. Kofler [3], W. Buchberger [2], S. Felber [1], F. Aichner [1,2]

Department of [1] Neurology, [2] Magnetic Resonance Imaging and Spectroscopy, University of Innsbruck, Austria, [3] Division of Restorative Neurology and Human Neurobiology, Baylor College of Medicine, Houston, Texas, USA

Key words: palatal myoclonus; olivary hypertrophy; head trauma; MR imaging

G. Birbamer, University of Innsbruck, Department of Neurology, Anichstraße 35, A-6020 Innsbruck, Austria.

Accepted for publication January 4, 1993

Palatal myoclonus, sometimes accompanied by other forms of segmental myoclonias, such as hyperkinetic movement disorders of the facial muscles, pharynx, larynx, diaphragm or extremities, is usually caused by a vascular ischaemic lesion of the brainstem or cerebellum (1, 2). Several other causes less frequently observed include encephalitis, degenerative diseases, multiple sclerosis, tumours and trauma (3–6). The characteristic pathological feature is hypertrophy of the inferior olivary nucleus (7–10). Recently, magnetic resonance (MR) imaging allowed direct visualization of inferior olivary hypertrophy associated with ischaemic or degenerative lesions in the brainstem or cerebellum (4, 11–14). MR findings in olivary hypertrophy due to severe traumatic brainstem injury have not yet been reported. We present three cases of traumatically induced olivary hypertrophy studied with MR-imaging.

Methods

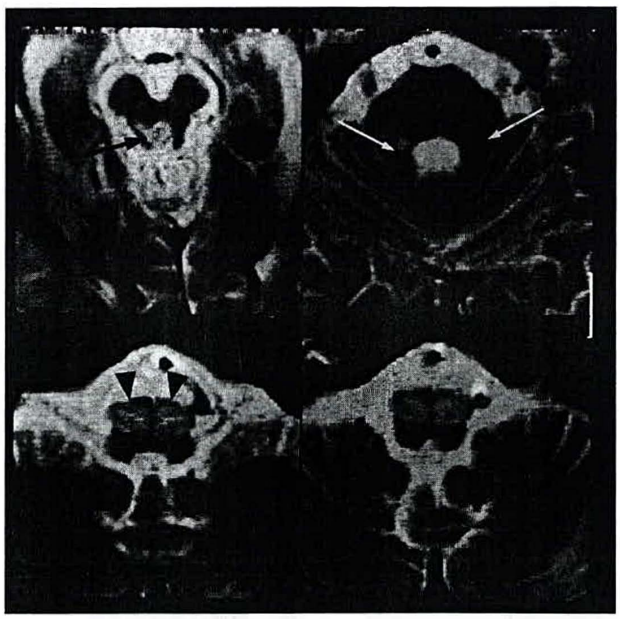
MR studies were performed on a 1.5 Tesla superconducting system (Magnetom, Siemens, Erlangen, Germany) using a circular polarized head coil (field of view = 25 cm). The imaging protocol consisted of sagittal T1-weighted images (TR = 500 ms, TE = 15 ms), axial and coronal proton density (PD) and T2-weighted images (TR = 2400 ms, TE = 15/90 ms). Additionally, a 3D flash sequence (TR = 40 ms, TE = 5 ms, flip angle = 40°) was applied. During imaging, the patients were monitored by continuous pulse oxymetry, capnography, ECG and blood pressure manometry.

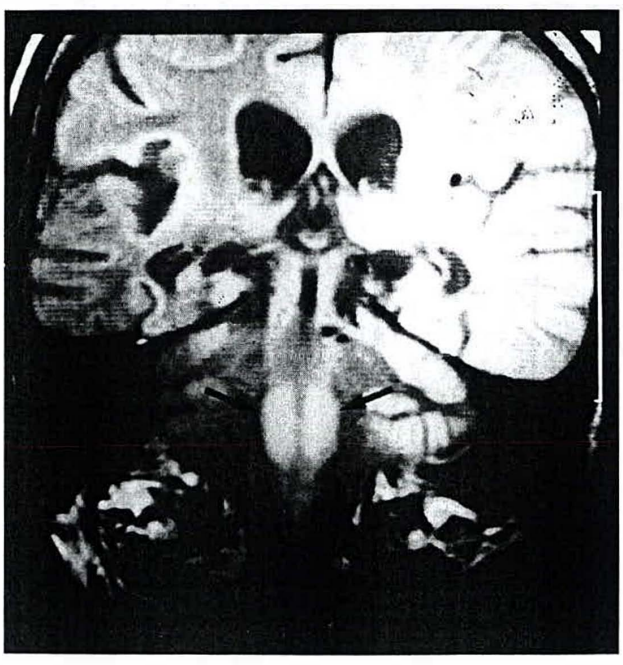
Case reports

Case 1. A 16-year-old boy sustained a severe head injury in a traffic accident. On admission he was comatose, asystolic and without spontaneous respiration. He required immediate cardiopulmonary resuscitation and artificial ventilation. The initial CT examination 2 h after the trauma, revealed diffuse brain edema. Four weeks later, CT showed signs of contusion in the mesencephalon, the splenium of the corpus callosum, and in the subcortical frontobasal and left frontoparietal white matter. The patient remained comatose and developed a traumatic apallic syndrome (15). Seventeen months after the injury, symmetric rhythmic contractions of the soft palate (palatal myoclonus) with a mean frequency of 120 min associated with rhythmic contractions of the pharynx and the perioral muscles and with intermittent upward-beating of the eyes were observed. In addition, the left arm and hand showed involuntary movements synchronously with the palatal myoclonus. All these involuntary rhythmic movements were continuously present during sleep and wakefulness. A right spastic hemiparesis was present, which might have prevented similar movements of the right upper extremity. The EEG showed no cor-

relation with the segmental myoclonic jerks. MR imaging 18 months after the injury revealed multiple lesions within the myoclonic triangle (Fig. 1 a–c). Areas of decreased signal intensity, suggesting local hemosiderin deposition, were present in the left upper cerebellar peduncle. The inferior olivary nuclei were enlarged symmetrically and showed increased signal intensity on PD-weighted and T2-weighted images. In addition, large areas of contusion were present bilaterally in the frontal lobe and thalamus, and in the splenium of the corpus callosum. Marked cortical and subcortical atrophy was noted.

Case 2. A 22-year-old man sustained a severe head injury in a motorcycle accident. On admission, he was comatose and presented with extensor posturing of the upper and lower extremities and with dilated pupils not reacting to light. Cerebral CT performed 3 h after the trauma, revealed multiple hypodensities, in the right temporal lobe, in both frontal lobes and in the left thalamus. Furthermore, subarachnoid hemorrhage and bleeding into the third and right lateral ventricle were observed. The patient remained comatose for four weeks and then developed a traumatic apallic syndrome. Clinically, spas-





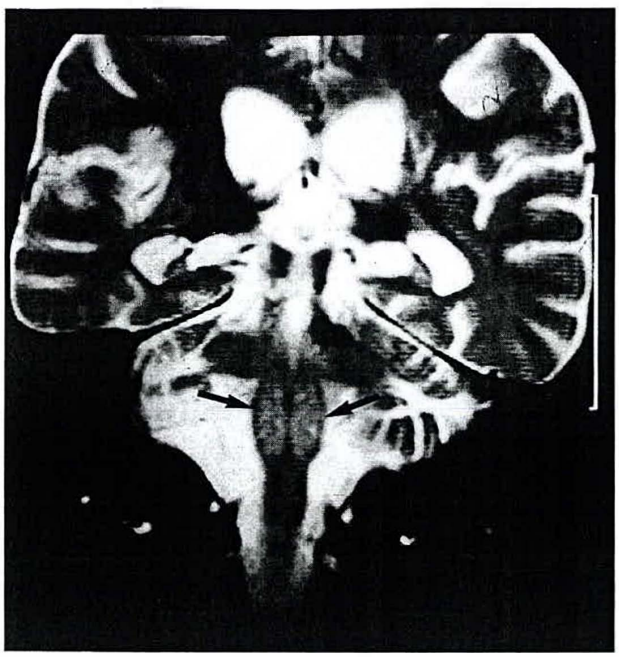


Fig. 1. (Case 1). (a) T2 weighted axial MR images showing areas of increased signal in the tegmentum (black arrows), posthemorrhagic lesions in both upper cerebellar peduncles (white arrows), and increased signal intensity and symmetrical enlargement of both inferior olivary nuclei (arrowheads); (b) T2 weighted coronal images showing symmetrical enlargement and increased signal intensity of both inferior olives (arrows); (c) PD weighted coronal images; the increase in signal intensity of the inferior olives is more prominent than in T2-weighted images (arrows).

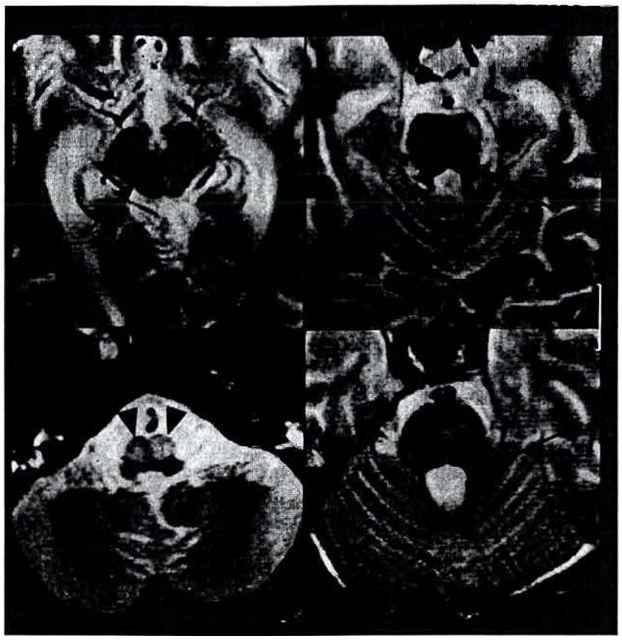


Fig. 2. (Case 2). T2-weighted axial MR images showing posthemorrhagic lesions in the tegmentum on the right side (arrow) and in the left upper cerebellar peduncle (arrow), and asymmetrical enlargement and increased signal intensity of the inferior olives (arrowheads).

tic tetraparesis more pronounced on the left side, gaze deviation to the left with sustained horizontal nystagmus, and right facial paresis were present. The clinical course was complicated by focal epileptic seizures, but no epileptiform discharges were found on interictal EEG recordings. During the following months, the patient's condition improved gradually, he responded to external stimuli and performed finalized skilful voluntary movements in all limbs. Ten months after the injury, palatal myoclonus and ocular bobbing appeared for the first time. MR imaging showed bilateral enlargement and increased signal intensity of the inferior olives on PD-weighted and T2-weighted images. The signal abnormality was more pronounced on the left side (Fig. 2). Areas of decreased signal intensity suggesting local hemosiderin deposition were shown in the left ventral and right lateral tegmentum next to the nucleus ruber. Posthemorrhagic lesions were also present in the left upper cerebellar peduncle and in the adjacent part of the vermis of the cerebellum. In addition, large cystic areas in both frontal lobes, consistent with resolving intracerebral hematomas, and signs of moderate atrophy were apparent in the brainstem and cerebellum.

Case 3. A 21-year-old patient sustained a severe head injury in a car accident. On admission, the patient was comatose and presented with a decortication pattern with flexor posturing of the upper and extensor posturing of the lower extremities, divergent position of the eye balls, and narrow pupils sluggishly reacting to light. The patient developed an apallic syndrome with poor remission during the following months. Cerebral CT 2 h after trauma showed diffuse cerebral edema and traumatic subarachnoid hemorrhage. MR imaging three months after the injury showed hemorrhagic contusions in the thalamic area and the splenium of the corpus callosum on the right side, the upper brainstem, involving the fibers of the central tegmental tract, and in the vermis and right dentate nucleus of the cerebellum.

Eighteen months after the injury, palatal myoclonus, vertical ocular myorhythmias ("upward beating nystagmus") and synchronous involuntary movements of the upper extremities ("wing beating") occurred. In addition to the earlier detected contusional lesions a repeat MR-examination showed bilateral enlargement of the inferior olivary nucleus with increased signal intensity on PD- and T2-weighted images.

Discussion

A variety of CNS structures, including the motor cortex, red nucleus, pretectal region, superior colliculi, vestibular nuclei, mesencephalic reticular formatio, contralateral cerebellar nuclei and the spinal cord, have connections with the inferior olivary nuclei (16). These connections terminate in a prominent dendritic pattern with abundant "boutons terminaux" (17). A lesion interrupting the central tegmental tract or dentato-olivary fibers from the dentate nucleus of the cerebellum may be associated with hypertrophic degeneration of the respective target neurons in the inferior olivary nucleus, resulting in an enlargement of the inferior olives (olivary hypertrophy) (2, 18).

Histopathologically, hypertrophic olivary degeneration is characterized by myelin loss vacuolar necrosis of neurons, and fibrillary gliosis (19, 20).

Guillain and Mollaret (3, 4) were the first to describe a relationship between lesions of the dentato-rubro-olivary pathway ("myoclonic triangle"), olivary hypertrophy, and the occurrence of segmental myoclonias. Their observations have been confirmed in several clinico-pathological studies (7, 8, 2, 5). Myoclonus of the soft palate is most commonly recognized, and is often accompanied by synchronous movements of the larynx, floor of the mouth, tongue, face, diaphragm, or skeletal muscles (2, 4, 8, 20, 21, 22, 23). Involvement of the oculomotor system is not rare, and the condition is then *termed* "oculopalatal myoclonus" (24). The combination of palatal myoclonus, vertical ocular myorhythmia and "wing beating" of the upper extremities has been observed in three patients with ischemic lesions of the brain-

stem (25). In our study, all patients had palatal myoclonus and vertical ocular myorhythmia, and two had synchronous unvoluntary movements of the upper extremities.

The time interval between the occurrence of a lesion within the dentato-rubro-olivary pathway and the clinical manifestation of palatal myoclonus is variable, although it peaks at 10 to 11 months (2). Our patients developed segmental myoclonias 11, 17 and 18 months after severe head injury. This delayed onset of symptoms may be explained by the development of denervation hypersensitivity of inferior olivary neurons. Thus, these neurons would become hyperactive in response to normal levels of circulating transmitters (23).

Segmental myoclonias have rarely been observed as a sequel of head injury (3, 26), but histopathologic confirmation of olivary hypertrophy was not available in these cases.

Recently, MR imaging has been used to study olivary hypertrophy and its associated lesions in the brainstem or cerebellum. Various reports described the MR appearance of olivery hypertrophy secondary to brainstem infarction, tegmental or cerebellar hemorrhage, and progressive ataxia (4, 11–14). Consistent with these observations, MR imaging showed bilateral enlargement of the inferior olives and increased signal intensity on PD weighted and T2-weighted images in all of our patients (Fig. 1a,b; Fig. 2). Bilateral involvement of the inferior olivary nuclei was probably due to the multilocularity of the traumatic lesions within the dentato-rubro-olivary pathway.

Generally, gliosis presents with increased signal intensity on T2-weighted images (27, 28). However, in all three cases the signal abnormality was more prominent on the proton density-weighted images (Fig. 1b,c). This suggests that the pathological changes in olivary hypertrophy are different from typical gliosis. High signal intensity on proton density-weighted images might be related to an increase in intracellular water content of enlarged neural bodies (13).

Based on our findings, we conclude that olivary hypertrophy can be detected by MR imaging in some patients following severe brainstem and/or cerebellar injury. The exact localization and neuroanatomical correlation of traumatic lesions by MR offers new insights to the patho-anatomical basis of different forms of segmental myoclonias.

References

1. Matsuo F, Ajax ET. Palatal myoclonus and denervation supersensitivity in the central nervous system. Ann Neurol 1979: 5: 72–78.
2. Lapresle J. Palatal myoclonus. Adv Neurol 1986: 43: 265–273.
3. Gerstenbrand F, Machacek F. Traumatisch bedingter Uvulanystagmus. Wien Z Nervenheilkunde 1968: 26: 293–303.
4. Sperling MR, Herrmann C Jr. Syndrome of palatal myoclonus and progressive ataxia: two cases with magnetic resonance imaging. Neurology 1987: 35: 1212–1214.
5. Jellinger K. Hypertrophy of the inferior olives. Report on 29 cases. Z Neurol 1973: 205: 153–174.
6. Hattori T, K Hirayama, Imai T, Yamada T, Kojima S. Pontine lesion in opsoclonus-myoclonus syndrome shown by MRI. J Neurol Neurosurg Psychiat 1988: 51: 1572–1575.
7. Kurachi M, Nakamura I, Katsukawa K, Kobayashi K, Sano Y, Isaki K, Yamaguchi N. Olivary hypertrophy in a case with palatal myoclonus: light- and electron-microscopic study. Folia Psych Neur JPN 1985: 39 (4): 543–550.
8. Gautier JC, Blackwood N. Enlargement of the inferior olivary nucleus in association with lesions of the central tegmental tract or dentate nucleus. Brain 1961: 84: 341–361.
9. Lhermitte J, Trelles JO. L'hypertrophie des olives bulbaires. Encephale 1933: 28: 588–600.
10. Vuia O. Aspects morphologiques (optiques et ultrastructuraux) de l'hypertrophie de l'olive bulbaire. Rev Neurol (Paris) 1976: 132: 51–61.
11. Hirono N, Kameyama M, Kobayashi Y, Udaka F, Mezaki T, Abe K, Nishitani N. MR demonstration of a unilateral olivary hypertrophy caused by pontine tegmental hematoma. Neuroradiol 1990: 32: 340–342.
12. Revel MP, Mann M, Brugieres P, Poirier J, Gaston A. MR appearance of hypertrophic olivary degeneration after contralateral cerebellar hemorrhage. AJNR 1991: 12: 71–72.
13. Yokota T, Hirashima F, Furukawa T, Tsukagoshi H, Yoshikawa H. MRI findings of inferior olives in palatal myoclonus. J Neurol 1989: 236: 115–116.
14. Zarranz J, Aurora F, Forcadas I. MR imaging of presumed olivary hypertrophy in palatal myoclonus. AJNR 1990: 1164.
15. Gerstenbrand F. Das traumatische apallische Syndrom. Springer Verlag, Wien, 1967.
16. Brodal A. Neurological anatomy in relation to clinical medicine. New York, Oxford: Oxford University Press 1981.
17. Scheibel ME, Scheibel AB. The inferior olive. J Comp Neurol 1955: 102: 77–131.
18. Goto N, Kaneko M. Olivary enlargement. Chronological and morphometric analyses. Acta Neurophathol (Berl) 1987: 54: 275–282.
19. Guillain G. The syndrome of synchronous and rhythmic palato-pharyngo-laryngo-oculo-diaphragmatic myoclonus. Proc R Soc Med 1938: 31: 1031–1038.
20. Guillain G, Mollaret P. Deux cas de myoclonies synchrones et rythmées velo-pharyngo-laryngo-oculo-diaphragmatiques. Le problème anatomique et physiopathologique de ce syndrome. Rev Neurol (Paris) 1931: 2: 545–566.
21. Deuschl G, Mischke G, Schenck E, Schulte-Mönting J, Lücking CH. Symptomatic and essential rhythmic palatal myoclonus. Brain 1990: 113: 1645–1672.
22. Trelles JO. Les myoclonies velo-palatines. Considérations anatomiques et physiopathologiques. Revue Neurologique 1968: 119: 165–171.
23. Dubinsky RM, Hallett M. Palatal myoclonus and facial involvement in other types of myoclonus. Advances in Neurology 1988: 49: 263–278.
24. Nakada T, Kwee TL. Oculopalatal myoclonus. Brain 1986: 109: 431–441.
25. Aichner F, Gerstenbrand F, Hengl W, Hollnsteiner W, Hoyt FW, Rumpl E. Über eine Kombination

von palatinem Myoklonus, regulärem vertikalen okulären My-orhythmen und "wing beating". In: GANNER H, ed. Referate der 15. Wissenschaftlichen Tagung der Gesellschaft Österre-ichischer Nervenärzte und Psychiater. Innsbruck, 1978: 46–47.
26. SHADDOCK SS, SHADDOCK LB, BLACK SPW: Palatal myoclonus following head injury. J Trauma 1972: 12: 353–357.
27. INOUE Y, MATSUMURA Y, FUDUKA T, et al. MR imaging of wallerian degeneration in the brainstem: temporal relationships. AJNR 1990: 11: 897–902
28. ORITA T, TSURUTANI T, IZUM HARA A, MATSUMAGA T. Coronal MR visualization of wallerian degeneration of the pyramidal tract. J Comp Ass Tomogr 1991: 15(5): 802–804.